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ABSTRACT

This poster reports on a study attempting to distinguish between the influences of domain-specific and domain-general developmental mechanisms in subjects with Williams Syndrome (a genetic defect resulting in mental retardation). Subjects included 10 monolingual French-speaking subjects (ages 9-21) with Williams syndrome and 10 monolingual French-speaking normal controls (5 years of age). It is argued that initial findings of the sparing of language in subjects with Williams syndrome have been premature, since this study shows both within- and across-domain dissociations for Williams syndrome. The comparison of different abnormal phenotypes and the identification of within-domain dissociations is seen as suggesting that some domain-specific predispositions exist, but that subsequent development calls on an intricate interplay of both domain-specific and domain-general mechanisms. (Contains 27 references.) (PB)

Within-domain dissociations in Williams syndrome: A window on the normal mind

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Abstract

In normal development, it is difficult to tease apart experimentally the influences of domain-specific versus domain-general mechanisms, yet such questions are central to developmental theorizing. We report a study of Williams syndrome which addresses this question. We show that the initial reports concerning the sparing of language in subjects with Williams syndrome are premature. Our experiments show both within- and across-domain dissociations for Williams syndrome. No theory that invokes only strong Nativism or only domain-general learning mechanisms applied across the board to different inputs can explain these dissociations. The comparison of different abnormal phenotypes and the identification of within-domain dissociations suggests that *some* domain-specific predispositions exist, but that subsequent development calls on an intricate interplay of both domain-specific and domain-general mechanisms.

WS neurobiological profile

==> 1 in 20,000 to 50,000 live births
==> Characteristic facial dismorphology
==> Renal and cardiovascular anomalies

Complete description of biological causes still unknown, but thought to involve:

==> defect in gene used to produce calcitonin and in gene used to produce the neurotransmitter calcium gene-related peptide (CGRP)

==> elevated calcium levels in WS blood possibly lead to elevated levels in brain during embriogenesis and/or post-natal growth, thereby upsetting timing/ location/use of calcium ions in brain cells

==> uniparental disomy has been ruled out, but it is possible that a micro-deletion, affecting a number of continugous genes, will be identified.

==> WS brain development results in volumetric abnormalities across different regions

WS Cognitive profile

==> IQs mainly in the 50-60s (range 40-90)

==> Verbal IQ usually outstrips Performance IQ

areas of DEFICIT

==> spatial cognition
==> number
==> problem solving

areas of PROFICIENCY

==> face recognition
==> theory of mind
==> language

Is WS language uniformly good?

In language tests, WS perform well on some structures (e.g. passive), but not on others (e.g. complex negatives). These selective difficulties, together with errors reported in parental questionnaires (e.g. grammatical gender), suggested the following:

that WS may be characterized not only by across-domain dissociations, but also by within-domain dissociations, e.g. within language itself.

As a preliminary exploration of within-domain dissociations, we tested a group of fluent-speaking French WS subjects on grammatical gender.

How grammatical gender works

Children must first learn article/noun pairs which have either masculine gender or feminine gender (e.g. un tapis [a carpet], une chaise [a chair]).

Gender is not based on semantics
(e.g. the word for "chair" [chaise] is feminine, "armchair" [fauteuil] is masculine; the two synonyms for "bicycle" [un velo; une bicyclette] are of different gender).

Gender can often be induced on the basis of word endings (e.g. -on is a typical masculine ending (e.g. un cochon [pig]), -onne [une couronne [crown]) is a typical feminine endings), but there are exceptions. However, this partially regular system based on word endings allows for generalization to novel words.

Earlier work had shown that normally developing children learn both the arbitrary article/noun pairs *and* induce the partially regular system.

Method

We used real words and nonce words to test generalization on the basis of either the article or the partially regular system of word endings.

Children were shown pairs of different coloured identical pictures. Colour terms were selected amongst those in French with audible gender marking:

e.g. vert/verte [green]
blanc/blanche [white]
gris/grise [grey]

Real words

Regular system:

e.g. un tapis, une flute.

Exceptions:

e.g. une fourmi, un parachute.

Child asked to name picture. Exp. then hid ring under one of the pair and asked where hidden. Child's response involved giving the article, noun and adjective:

Exp: "J'ai caché ma bague ...?
[I hid my ring...?]

Child: "sous le tapis vert"
"sous la fourmi verte", etc.
[under..]

Nonce terms

Regular system:
e.g. un bicron, une spodine.
Exceptions:
e.g. une plichon, un forsine.

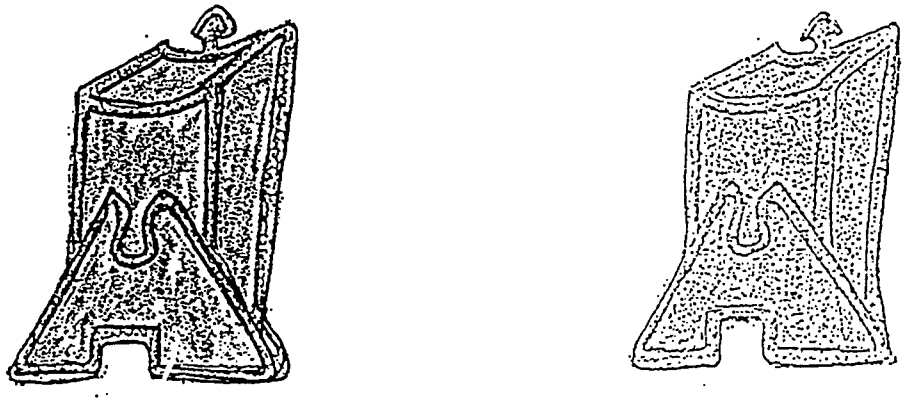
Exp. showed child pictures of strange objects/animals and named them. Then the child repeated the name. In some conditions we provided the article obeying the system (e.g. ça c'est une coumette [that's a coumette], or violating it ("ça c'est une plichon [that's a plichon]). In other conditions we provided only the ending (e.g. ça c'est deux mattons [that's two mattons]. Here the child must induce gender from the ending alone. Then E. hid ring under one picture and asked:

Exp: "J'ai caché ma bague ...?
Child: "sous la coumette blanche"
"sous le plichon blanc"

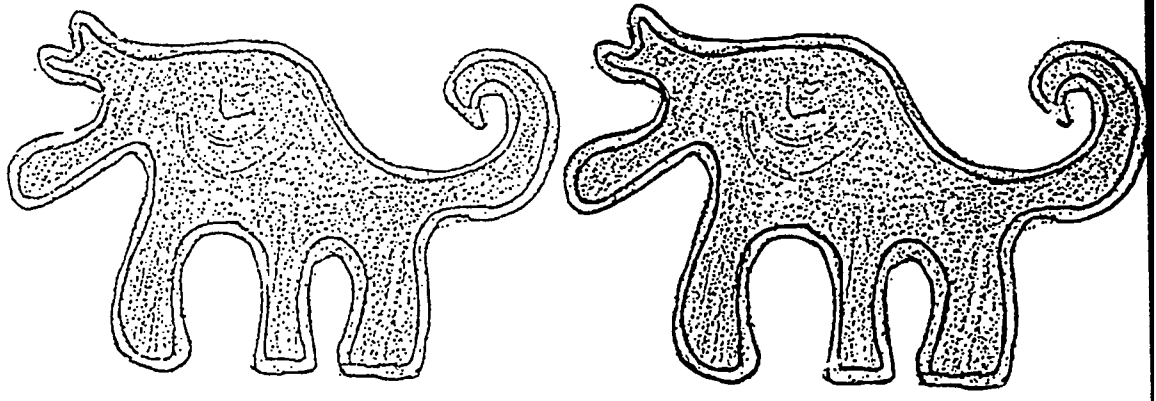
Typical pairs of coloured stimuli for nonce terms

green [vert/verte]
grey [gris/grise]

objects



animals



Population

10 monolingual French-speaking WS subjects

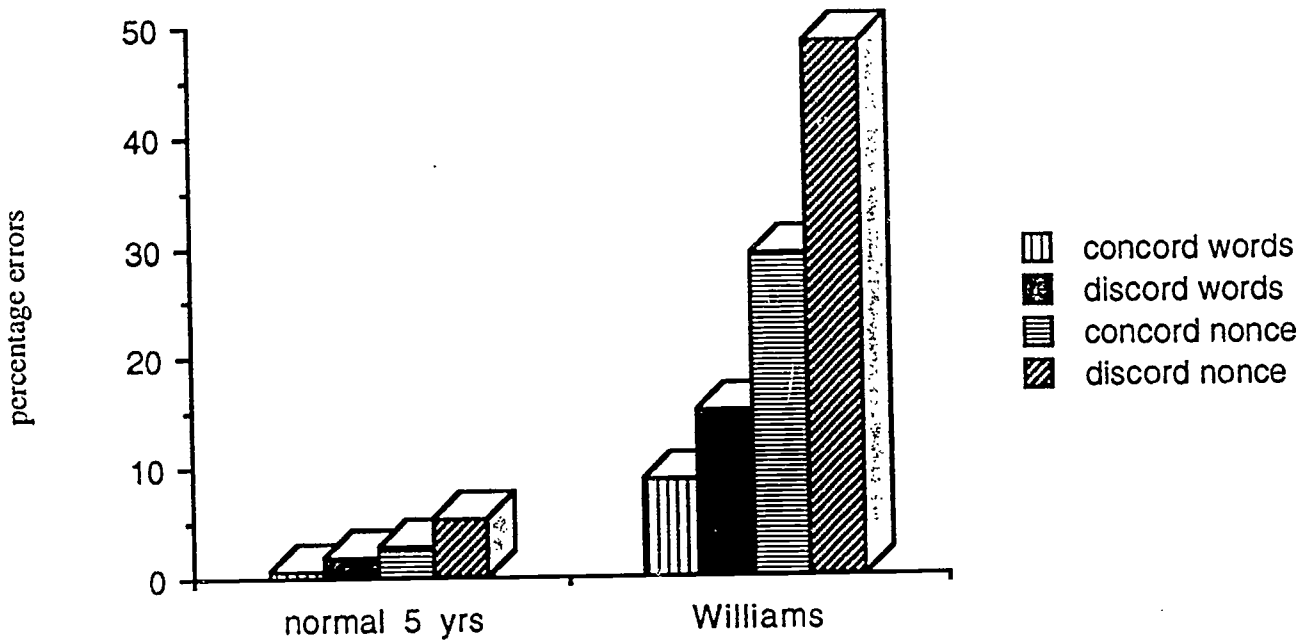
Chronological age: 9 - 21 years
IQ range: 51 - 67

10 monolingual French-speaking normal controls

Chronological age: 5,1 - 5,10 years

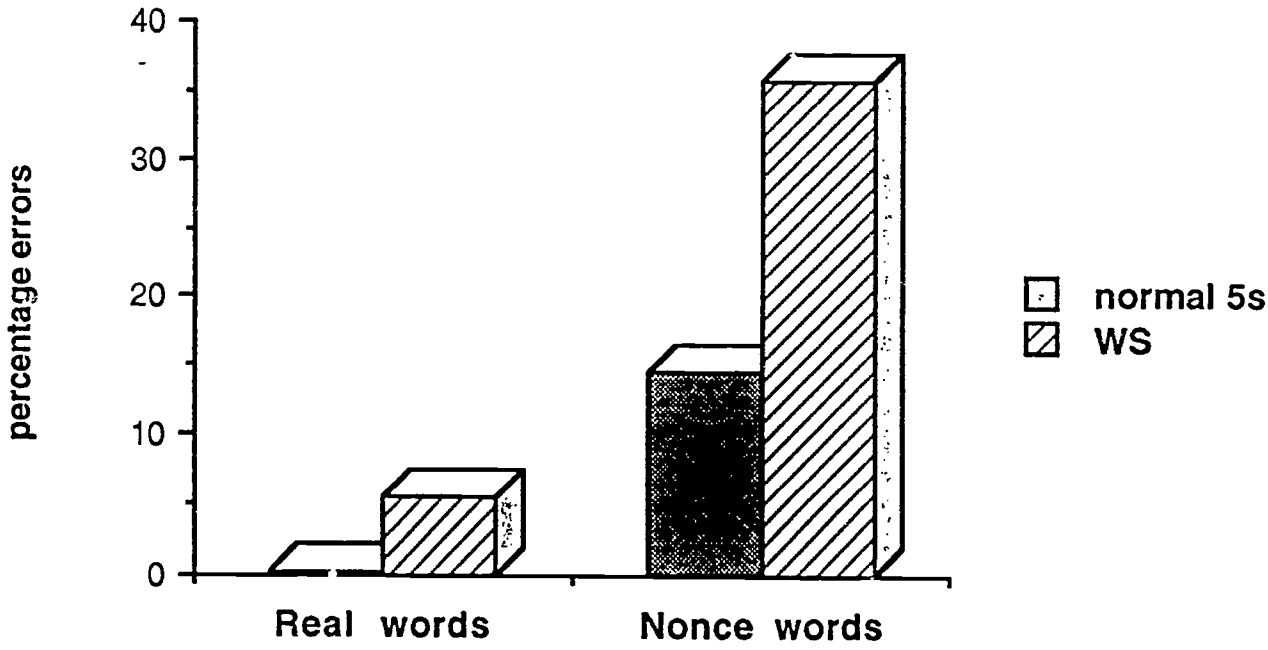
Results

N.B. General pattern of errors across the different categories of stimuli is similar for both populations: real words are easier than nonce words, items with concord between article and ending are easier than those with discord. The larger number of errors for WS subjects suggests that they only weakly extract the underlying system.



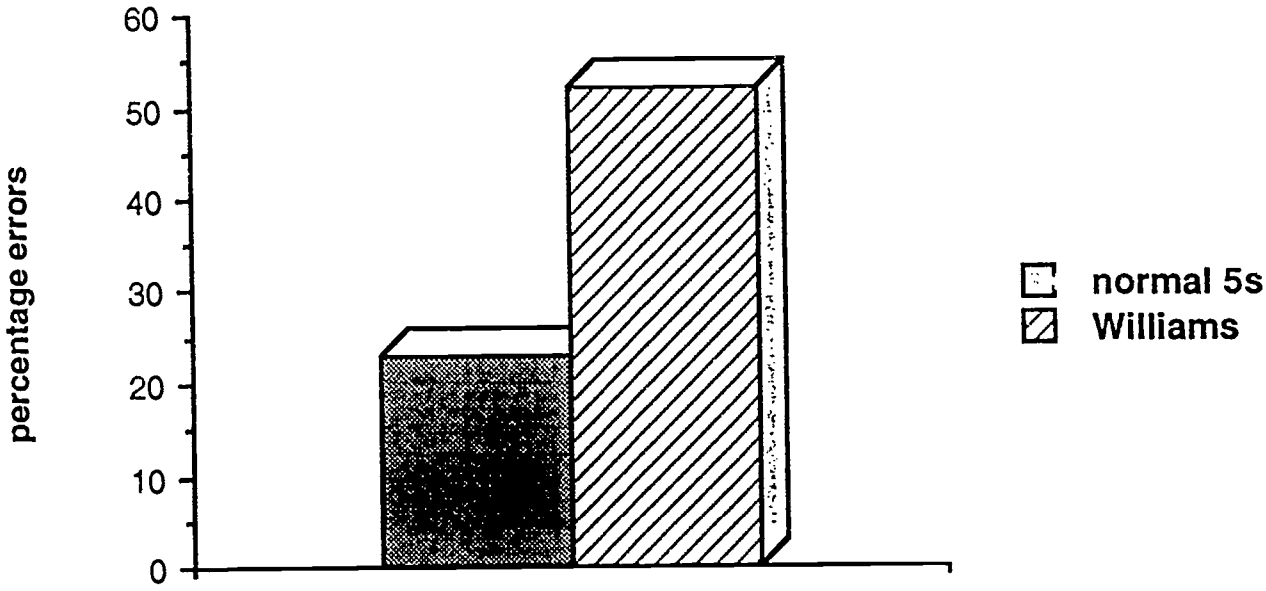
Failure to make adjectives agree with cues on articles

(e.g. given "une plichon", child replies "la plichon vert", instead of "la plichon verte")



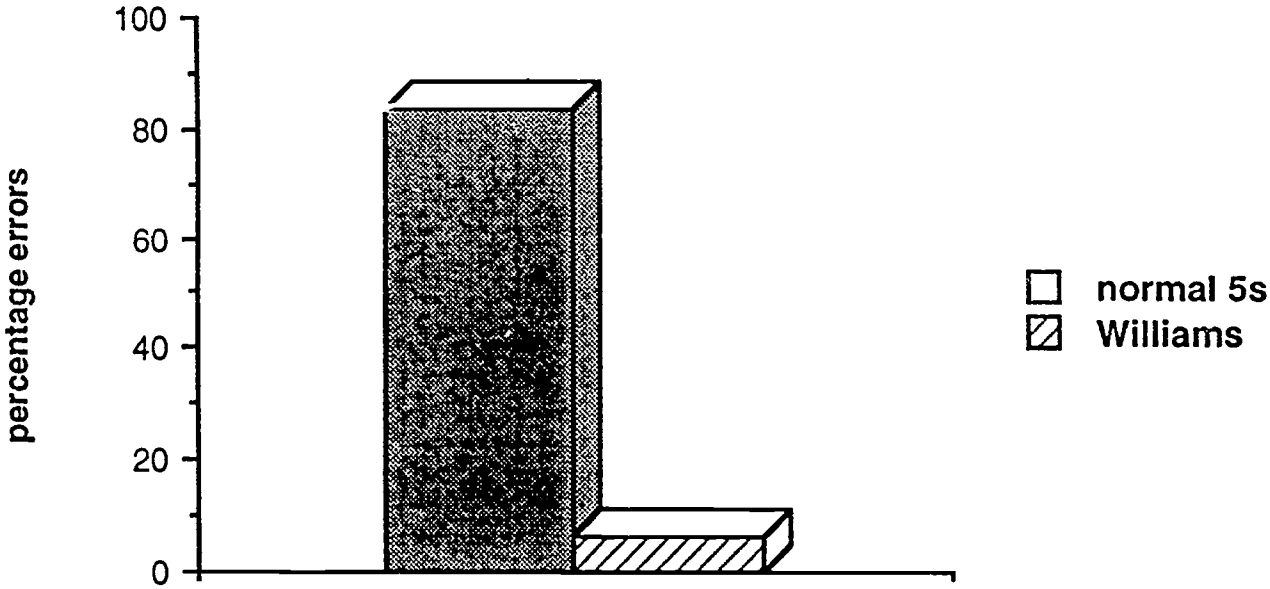
Failure to make use of cues on endings

(e.g. given "deux faldines", child replies "le faldine", instead of "la faldine")



Failure to repeat nonce words correctly on first attempt

(e.g. given "bicron", child repeats "bitonne")



N.B. WS perform better than normal controls

Discussion

Why do WS subjects perform so poorly on gender tasks?

==> *Not* because they have difficulty with formal test situations

==> *Not* because they cannot recall the nonce terms - in fact they recall them better than controls

==> *Not* because they do not know the masculine and feminine forms of articles and adjectives

==> *Not* because they are generally poor at language production

But because of several possible reasons:

==> normal children learn grammatical gender by simultaneously acquiring exemplars of article/noun pairs by rote *and* by extracting the generative system of oppositions on word endings; they are good system and theory builders.

==> WS children are good at learning exemplars of article/noun pairs by rote via domain-specific mechanisms, but their domain-general limitations mean that they only weakly extract the underlying system of oppositions on word endings. They tend to be poor at system and theory building.

==> It is also possible that arbitrary systems like gender have to be learnt very early in the acquisition process and it is known that WS children are often delayed in their acquisition - this suggests that there may be critical learning periods for different aspects of language.

==> Studies of vocabulary acquisition show that WS are less sensitive to frequency than normal subjects. The partial system of morpho-phonological oppositions on word endings for French gender is crucially dependent on input frequency.

General Implications

==> Focus on a single syndrome or on global domain-specific analyses can lead to missing the differences between uneven cognitive profiles or failing to discover the existence of within-domain dissociations, thereby leading to erroneous generalizations:

e.g. Down syndrome has been used to invoke a domain-general explanation of development and its impairments

e.g. Williams syndrome has been used to invoke a general language module

==> If a single learning mechanism were used to learn all aspects of language, as some connectionist models propose, then why do WS find grammatical gender so problematic and much of syntax so easy?

==> Our results suggest that we need to invoke, not one, but multiple learning mechanisms which are brought to bear on language acquisition, e.g.

==> one dealing with domain-specific syntactic mapping

==> one dealing with lexical acquisition

==> one calling on domain-general inductive mechanisms (weak in WS subjects) to generate productive systems (like arbitrary grammatical gender marking) which cannot be bootstrapped by semantic or syntactic processors

References

Bellugi, U., Marks, S., Bihrle, A.M. and Sabo, H. (1988). Dissociation between language and cognitive functions in Williams Syndrome. In D. Bishop & K. Mogford, (Eds.), Language Development in Exceptional Circumstances. London: Churchill Livingstone.

Bellugi, U., Bihrle, A., Neville, H., Jernigan, T. & Doherty, S. (1992) Language, Cognition and Brain Organization in a Nuerodevelopmental Disorder. In M.Gunnar & C.Nelson (Eds.) Developmental Behavioral Neuroscience. Hillsdale, NJ: Lawrence Erlbaum Associates.

Bellugi, U., Wang, P. & Jernigan, T.L. (in press) Higher cortical functions: evidence from specific genetically based syndromes of disorder. In S.Broman & J. Graffman (Eds.) Cognitive Deficits in Developmental Disorders: Implications for Brain Function. Hillsdale, NJ: Lawrence Erlbaum Associates.

Bihrle, A.M., Bellugi, U., Delis, D., & Marks, S. (1989) Seeing either the forest or the trees: Dissociation in visuospatial processing. Brain and Cognition, 11, 37-49.

Carey, S. (1993) Poster on the development of biological theories in children with Williams syndrome. SRCD, New Orleans.

Culler, F., Jones, K., & Deftos, L. (1985). Impaired calcitonin secretion in patients with Williams syndrome. Journal of Pediatrics, 107, 720-723.

Donnai, D. & Read, A.P. (1993) Imporinting, uniparental disomy and Williams syndrome. Final Report to The Infantile Hypercalcaemia Foundation, U.K.

Jernigan, T.L. and Bellugi, U. (1982) Neuroanatomical distinctions between Williams and Down syndromes. In S.Broman & J.Grafman (Eds.) Atypical Cognitive Deficits in Developmental Disorders: Implications for Brain Function. Hillsdale, NJ: Laurence Erlbaum Associates.

Jernigan, T.L. and Bellugi, U. (1990) Anomolous brain morphology on Magnetic Resonoance Imaging in Williams and Down syndrome. Archives of Neurology, 47, 529-533.

Jernigan, T.L., Bellugi, U., Sowell, E., Doherty, S. & Hesselink, J.R. (in press) Cerebral morphological distinctions between Williams and Down syndromes. Archives of Neurology.

Johnson, M.H. and Karmiloff-Smith, A. (1992) Can neural selectionism be applied to cognitive development and its disorders? New Ideas in Psychology.

Johnson, M.H. and Morton, J. (1991). Biology and Cognitive Development: The case of face recognition. Oxford: Blackwell.

Karmiloff-Smith, A. (1990) Piaget and Chomsky on language acquisition: divorce or marriage? First Language, 10, 255-270.

Karmiloff-Smith, A. (1992a) Beyond Modularity: A developmental perspective on cognitive science. Cambridge, MA: MIT Press/Bradford Books.

Karmiloff-Smith, A. (1992b). Self-organization and cognitive change. In M.H.Johnson (Ed.) Brain Development and Cognition: A reader. Oxford: Blackwells.

Karmiloff-Smith, A. (1992c) Abnormal phenotypes and the challenges they pose to connectionist models of development. Technical Report Series on Parallel Distributed Processing and Cognitive Neuroscience, No. PDP/CNS.92.7, Carnegie Mellon University, Pittsburgh.

McKusick, V. (1988) Mendelian inheritance in Man: Catalogs of autosomal dominant, autosomal recessive and X-linked phenotypes. Baltimore: Johns Hopkins Press.

Marriage, J. (in progress) The developmental pattern of hyperacusis in children with Williams syndrome.

Mervis, C. and Bertrand, J. (1993) General and specific relations between early language and early cognitive development. Paper presented to the Biennial Meeting of the SRCD, New Orleans, March 1993.

Morris, C., Demsey, S., Leonard, C., Dilts, C. & Blackburn, B. (1988) Natural history of Williams syndrome: Physical characteristics. Journal of Pediatrics., 113, 318-326.

Murphy, M.B., Greenberg, F., Wilson, G., Hughes, M. & DiLiberti, J. (1990) American Journal of Medical Genetics Supplement, 6, 97-99.

Neville, H.J. (1991). Neurobiology of cognitive and language processing: Effects of early experience. In K.R. Gibson and A.C. Petersen, (Eds.), Brain Maturation and Cognitive Development: Comparative and cross-cultural perspectives. Hawthorne, NY: Aldine deGruyter Press.

Price, J.L., Russchen, F.T., & Amnaral, D.G. (1987) The amygdaloid complex. In L.W. Swanson, A.Bjorklund & T.Hokfelt (Eds.) Handbook of chemical neuroanatomy. Vol.5, , (pp.279-388). New York: Elsevier.

Udwin, O. (1990) A survey of adults with Williams syndrome and idiopathic infantile hypercalcaemia. Developmental Medicine and Child Neurology, 32, 129-141.

Udwin, O., Yule, W. and Martin, N. (1987). Cognitive abilities and behavioural characteristics of children with ideopathic infantile hypercalcaemia. Journal of Child Psychology and Psychiatry, 28, 297-309.

Udwin, O., and Yule, W. (1991) A cognitive and behavioural phenotype in Williams Syndrome. Journal of Clinical and Experimental Neuropsychology, 13, 2, 232-244.

Wang, P.P.., Hesselink, J.R. Jernigan, T.L., Doherty, S. & Bellugi, U. (in press) The specific neurobehavioral profile of WS is associated with neocerebellar hemispheric preservation. Neurology.